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Discussion

Dr W. Williams (Toronto, Ontario, Canada). Mr. Chairman, I have no conflicts of interest to disclose.

I would like to congratulate Dr Shin'oka and his colleagues for their report of their excellent, very large clinical experience in surgical management of patients with AV discordance. Their analysis focuses on survival and the factors that affect survival. Specifically, they address the important question of whether these patients are better served by an anatomic repair wherein the left ventricle is connected to the systemic circulation, rather than conventional repair in which the morphologic right ventricle remains the systemic ventricle. It's generally accepted that the right ventricle and its tricuspid valve are poorly suited to support the systemic circulation lifelong. There are, however, rare cases of isolated AV discordance in patients who have no associated lesions and who survive into the seventh and eighth decade of life, perhaps the exceptions that prove that rule.

The majority of patients with AV discordance have major associated lesions. The most prevalent are VSD, PS, or PA, TR, and AV block.

These 4 lesions occur in permutations and combinations, but, of course, any cardiac lesion may occur with AV discordance including single ventricle. In Dr Shin'oka's series, 21% of their patients had a Fontan operation for single ventricle.

In 1990, Dr Ilbawi reported success with anatomic repair for AV discordance as an alternative to conventional repair. Ilbawi's contribution of using the left ventricle in the systemic circulation expanded the options for these patients. There are now at least 8 surgical options to manage patients with AV discordance. Whether the more complex anatomic repairs will produce better long-term results remain unknown. I must say I am concerned about combining, in one patient, the well-known late complications of an atrial repair, whether a Mustard or a Senning, and those of the Rastelli-type repair.

Because there are so many surgical options and AV discordance is a rare lesion in which there are 4 commonly associated lesions that occur in various combinations, it is not surprising that the Tokyo series failed to demonstrate a difference in long-term outcome whether the left ventricle or the right ventricle was connected to the systemic circulation.

Indeed, the survival of their single-ventricle Fontan patients was identical to that of either anatomic or conventional repair.

Dr Shin'oka is not alone in failing to demonstrate a difference in survival for these various surgical approaches. My colleagues, Dr Brian McCrindle and Dr Glen Van Arsdel, recently published a meta-analysis of AV discordance in which they reviewed more than 60 papers. Their conclusion is worth repeating: "it will require a well designed, large, multicenter cohort study as the only practical solution in resolving the optimal choice of surgical procedures for these patients."

I have two questions for Dr Shin'oka. Number one, in the manuscript and in the bar graph that you showed, conventional repair was used during the most recent era in about 30% of the patients. Does this recent experience suggest a trend toward return to conventional repair? What are your current indications for conventional repair?

Dr T. Shin'oka (Tokyo, Japan). Thank you, Dr Williams, for your thoughtful comments and question. As you stated in your comments, I do agree that we need a large multicenter study to determine the optimal choice of surgical procedure for this rare entity using a more sensitive end point.

In response to the first question, after 2000, we have demonstrated a trend toward a return to conventional repair when a patient has a relatively small-sized VSD and requires the VSD enlargement with a Rastelli procedure. In addition, VSD enlargement was a risk factor for the PM implantation, although VSD enlargement was not a risk factor for HD or LD.

Dr Williams. My second question is, what are your current indications for preparation of the left ventricle by pulmonary artery banding? In the series you reported today, 7 of the 15 patients had a PA band prior to a DSO. Other patients had PA bands but did not undergo anatomic repair. Given your considerable experience, which patients would you currently select for a PA band and in whom would you not attempt a PA band for preparation of the left ventricle?

Dr Shin'oka. This question concerns the indication of pulmonary artery banding for RV training. In our series, 7 of 15 patients underwent pulmonary artery banding prior to the arterial switch operation. The other 8 nonbanded patients had high LV pressure due to the VSD or subpulmonary stenosis; therefore, we did not have to do the banding before the switch operation. Also, 4 of the 7 banded patients had VSD, and 3 of the 7 banded patients had an intact ventricular septum. These 3 patients with an intact ventricular septum required LV training before the arterial switch operation. They also had very severe TR and RV failure before PA banding. In our country, heart transplantation is very limited. Therefore, in such a case, we attempt a PA banding procedure when the patient's family elects not to have direct tricuspid valve surgery after we explained the results of our past series.

Actually, selecting the best procedure is difficult in these patients with an intact ventricle septum and severe TR.

Dr C. Tchervenkov (Montreal, Quebec, Canada). I'd like to ask the question, as the long-term survival doesn't appear to be different between these various options, then we ought to look for additional benefits that the patient may have from these more complex anatomic repair procedures. Do you have any data on the functional assessment and status of these patient in terms of quality of life, exercise tolerance, et cetera, to help us elucidate which is the best option?

Dr Shin'oka. I agree with you. As Dr Williams pointed out, we need a large multicenter study to determine the optimal choice of surgical procedure for this rare entity using a more sensitive end point. We are currently analyzing the precise function of the anatomic left ventricle or right ventricle in a long-term period. We may be able to present this precise data at the next meeting.

Dr G. Van Arsdel (Toronto, Ontario, Canada). I would just add to that. It would be helpful to see VO_2 data on these patients to understand whether or not we are achieving functional improvement. You have a significant cohort on whom you could perform that study.

In the meta-analysis we published last month, we noted that in the more recent era there was actually early survival benefit to doing an anatomic-type Rastelli–Mustard repair as opposed to a physiologic repair. That may be because of 2 reasons: 1, when you do a physiologic repair, TR may be an immediate consequence; and 2, there is an immediate demand for the morphologic right ventricle to carry the systemic load without the benefit of a morphologic LV assist (closed VSD).

Your survival data appear to demonstrate this phenomenon. It's true at later follow-up the Rastelli-type repair and the physiologic repair appeared to be the same, but early outcome at 1 to 2 years seemed to be different. Did you divide your time of follow-up in outcomes to see whether or not there was a difference?

Dr Shin'oka. Thank you, Dr Arsdel, for your careful observation of our survival curve. We did not perform such an analysis, but that analysis may be useful for the further evaluation.

Dr Van Arsdel. The second question I had, I was interested in the fact that you would accept a VSD 50% of the size of the aorta before you would enlarge it. But there have been some data to suggest that VSD enlargement impairs ventricular function, and we've actually felt that if you need to enlarge the VSD, perhaps we should do a more conventional-type repair. Were you able to follow the ventricular function on those patients who had a VSD enlargement and was it a predictor of poor ventricular function or poor outcome?

Dr Shin'oka. In the patients who underwent the Rastelli–Senning-type procedure, 33 of 69 patients had a VSD enlargement. Although I did not show the data, we compared LV function after surgery between the VSD-enlarged group and the nonenlarged group. The LV ejection fraction after surgery in the VSD enlarged group was 54.3%, whereas the LV ejection fraction after surgery in the nonenlarged group was 53.2%. We did not find any statistical difference between the groups.

Dr A. Corno (Liverpool, United Kingdom). You have used a long series of different surgical techniques, including, more or less, all the types of biventricular repair and the univentricular type of repair, but you have never used the one-and-a-half type of ventricular repair, consisting of arterial switch or Rastelli, the atrial rerouting limited to the inferior vena cava associated with bidirectional Glenn.

This option presents several advantages: (1) you have no risk of superior vena cava obstruction; (2) you increase the intra-atrial space available for the pulmonary venous return, therefore you reduce the risk of obstruction to the pulmonary venous return; (3) you reduce the intra-atrial suture lines, so you reduce the risk of arrhythmias; (4) you unload the right ventricle, and this is very useful when you have a relative hypoplasia or malfunction of the right ventricle; (5) you reduce the duration of the ischemic time; and (6) you also reduce the flow through the right ventricle to pulmonary artery connection.

My question is the following: in the patients from your study that are now considered, based on your experience, at higher risk, would you consider this option in the future?

Dr Shin'oka. Thank you, Dr Corno. Yes, I agree with the other option you recommended. We would like to try this option in the future either in conventional or anatomic repair when the patients have a relatively small-sized pulmonary ventricle.

Dr G. Stellin (Padova, Italy). You showed that the complete AV block is a frequent complication in repairing corrected transposition and especially when a VSD needs to be enlarged. You showed in your drawing that the incision is carried posteriorly, in the VSD. Do you use the same approach in IDD-corrected transposition?

Dr Shin'oka. Most patients with IDD also had an anterior conduction system in our series, and we performed the electrophysiologic studies before the operation. This study can determine

the dominant AV node in almost all patients. Therefore, according to these findings, we can enlarge the VSD correctly.

Dr Stellin. According to the anatomist, the conduction system goes with a loop, so if you have an L loop, it's anterior. If you have a D loop, it's posterior. So if you have a large atrial septal defect in an IDD form or corrected transposition, you might indeed have injured your His bundle.

Dr Shin'oka. I do not agree with your opinion. It's not correct. There is a general agreement of an anterior conduction system in

the situs solitus group, and a posterior conduction system was reported in the situs inversus group. However, we have observed many patients with an anterior conduction system in IDD, 16 of 21 patients. In addition, a dominant posterior conduction system was noticed even in an SLL heart, in 5 of 48 patients. Furthermore, a sling of conduction bundle was reported in cc-TGA in situs solitus with a straddling mitral valve. Therefore, the direction of the VSD enlargement should be carefully determined by means of the preoperative electrophysiologic study.